Lipoedema- a fat disorder: considerations for General Practitioners.

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Abstract

Lipoedema is a fat and connective tissue disorder associated with proliferation of diet-resistant inflammatory fat tissue. It predominately affects women and commonly manifests as symmetrical enlargement of the legs, buttocks and arms. Lipoedema often develops or worsens at key stages of life such as puberty, pregnancy and menopause, and is associated with a variety of physical and psychosocial problems. It is poorly understood and women may be wrongly diagnosed as having lymphoedema or 'obesity', leading to delay in them accessing appropriate treatment and support. Lipoedema aetiology appears to include genetic, immune and endocrine factors, although the science is evolving, research is ongoing and, in the future, a clearer picture of lipoedema pathophysiology will emerge. This paper examines evidence relating to lipoedema pathophysiology, diagnosis, and the psychological impact of lipoedema, providing an overview of current treatment and self-management approaches, and key considerations for GPs.

Keywords: lipoedema; secondary lymphoedema; pathophysiology; psychosocial distress; self-management; liposuction.

Introduction

Lipoedema is a fat and connective tissue disorder that predominately affects women and commonly manifests as symmetrical enlargement of the legs, buttocks and arms (Fife et al 2010). Although due to a proliferation of diet-resistant inflammatory fat tissue, lipoedema is a poorly understood condition, often wrongly diagnosed as lymphoedema or 'obesity' (Langendoen et al 2009). Lipoedema aetiology appears to include genetic, immune and endocrine factors (Szél et al 2014; Wounds UK 2017), although research evidence regarding pathophysiology is still limited and evolving. Epidemiological research has estimated the prevalence of lipoedema in the UK to be 1 in 72,000 population (Child et al 2010), a figure that is likely to be underestimated (Wounds UK 2017).

Lipoedema can have a significant influence on quality of life. It often develops or worsens at key stages of life such as puberty, pregnancy and menopause, and is associated with a variety of physical and psychosocial problems (Dudek et al 2016; Dudek et al 2018; Fetzer & Fetzer 2016). This paper discusses current evidence relating to lipoedema pathophysiology, diagnosis, and the psychological impact of lipoedema. It provides an overview of current treatment and self-management approaches, drawing on a UK Best Practice Document Wounds UK 2017), and key considerations for general practice nurses (GPNs). Examples of patient stories related to reproductive health issues in women with lipoedema are also presented.

Lipoedema pathophysiology and features

This is likely to be multi-factorial but it appears to involve adipocyte hypertrophy (increase in the size of fat cells) and hyperplasia (increased numbers of fat cells), with proliferation of adipose-derived stem cells and progenitor cells in the tissues (Suga et al 2009). It is hypothesised that increased adipogenesis and tissue enlargement may trigger hypoxia, adipocyte death, and infiltration of the tissues by inflammatory products including macrophages (Okhovat and Alavi 2015). Notably, there is reduced elasticity of the skin and connective tissues, which may contribute to the development of fat pads medial to the knee, particularly characteristic of late stage lipoedema (Table 1).

Other features of lipoedema include easy bruising, possibly due to the growth of fragile blood capillaries which may leak, and contribute to the development of

secondary lymphoedema, when the lymphatic system becomes overwhelmed. There are some reports of changes in the structure and function of the lymphatics in lipoedema, such as slowing of lymphatic flow (Bilancini et al 1995; Wounds UK 2017). Pain may also be related to joint problems, particularly the knees and hips and many women with lipoedema describe problems with hypermobility indicating some degree of tissue laxity (Wounds UK, 2017).

Pain is a primary complaint in women with lipoedema (Romeijn et al 2018), usually when local pressure is applied to the lipoedematous area, possibly due to compression of nerve fibres due to inflammation or central sensitisation, similar to processes in chronic pain (Wounds UK 2017). Cellulitis can be a complication of later stage lipoedema, when lymphoedema is present or can also develop after an event such as an insect bite on the lipoedematous area (Williams & MacEwan 2017).

Table 1: Stages of lipoedema with signs and symptoms (Forner- cordero et al 2009; Peled and Kappas 2016; Wounds UK 2017)

Stage of lipoedema	**Description**	**Characteristic signs and symptoms on clinical examination**
Stage 1	Early bilateral and symmetrical enlargement, mainly of the lower limbs, but may include buttocks and arms	• Skin is smooth, soft and may feel cool • The feet are not enlarged • 'Inverse shouldering' occurs at the ankle (ankle cuff) • Easy bruising may occur • Stemmer's sign is negative • Pain on gentle pressure to the lipoedema areas
Stage 2	The skin texture becomes irregular and indurated and tissue enlargement extends; may include buttocks and abdomen	• As above • Indurations of the fat tissue develop • Subcutaneous nodules may be palpated in the tissues
Stage 3	Fat tissue indurations extend, and deformed lobular fat pads begin to develop particularly around the medial thigh and knees	• The soft fatty tissues fold in on themselves and limb shape becomes distorted by the fat pads • Subcutaneous nodules or lipomas may be more obvious • Skin may have the texture of orange peel or be dimpled • Fat pads lead to abnormal gait • Muscle weakness may be evident

		• Stemmer's sign is usually negative but interstitial fluid may lead to pitting oedema • Pain on pressure to the lipoedema tissue, and hip/knee joint pain
Stage 4	Secondary lymphoedema occurs sometimes referred to as 'lipo-lymphoedema) due to the lymphatic system being overwhelmed	• Stemmer's sign may be positive • The feet and hands may be involved • Signs of protein-rich lymphoedema occur including persistent swelling, local hyperkeratosis, lymphangiectasia, papillomatosis, lipodermatosclerosis, cellulitis • Swelling and fibrosis may be particularly evident in the fat pads, abdominal apron and distal limbs • Mobility and function may be significantly affected due to distorted limb shape, and muscle weakness (Smeenge 2013).

Diagnosis of lipoedema

At present there is no specific diagnostic marker for lipoedema, and diagnosis is made on history, clinical examination, and identification of characteristic signs and symptoms (Table 2). It is common to hear that the woman first noticed swelling in her teenage years, became focussed on dieting, which was unsuccessful despite careful adherence to a variety of diets. Further questioning may elicit information about family members who also appear/ed to have lipoedema, often undiagnosed, but evident in family photos. Women may report significant changes in lipoedema with subsequent pregnancies, at menopause, during hot weather, when travelling, standing or sitting for long periods.

Although lipoedema must be differentiated from obesity, some women experience additional problems with weight changes due to an increase in non-lipoedema fat, particularly if mobility or psychological wellbeing is compromised by the lipoedema (Fife, 2010). Importantly, the disproportionate (pear) body shape of lipoedema may be obvious where the upper body is much smaller than the lower; this readily differentiates lipoedema and 'obesity' (Buck and Herbst, 2016; Peled and Kappos, 2016).

Table 2: Diagnostic considerations in lipoedema (Wounds UK 2017)
• History and examination to identify characteristic changes that enable differential diagnosis • Exclude/investigate for other conditions that contribute to fat changes/weight gain/oedema, including organ failure (Peled & Kappos 2016), and hormonal conditions • Consider presence of co-existing conditions including hypermobility, muscle weakness, hip and knee joint changes and osteoarthritis, allergies and gut sensitivities • Consider presence of psychological distress, poor mental health or disordered eating that may require additional support and influence adherence to self-care • Consider inflammatory markers such as C-reactive protein if cellulitis is suspected • Avoid undue focus on weight gain and loss; body mass index is a limited tool in lipoedema • Referral to vascular to exclude treatable venous disease if indicated • Lymphoscintigraphy is rarely indicated but may be occasionally used to identify lymphatic abnormalities.

Psychological impact of lipoedema

A number of surveys have provided insights into the impact of lipoedema on women. In the UK, an online survey undertaken between 2012 and 2014 by Lipoedema UK (Fetzer & Fetzer 2016) reported on the experiences of 250 women with lipoedema. The average age of lipoedema diagnosis was 44 years, with many reporting significant experience of misdiagnosis over many years and 87% reporting lipoedema as having a negative effect on their quality of life. Furthermore:

- 98% had tried to lose weight
- 76% had difficulty in losing weight
- 95% had difficulty with buying clothes
- 51% found lipoedema had affected their career

- 60% reported a restricted social life
- 50% reported a restricted sex life
- 45% reported experiencing an eating disorder.

Romeijn et al (2018) undertook an email survey of 163 Dutch women with lipoedema, incorporating questionnaires including the RAND-36. They reported the average time from onset of lipoedema to diagnosis as 18 years, identifying quality of life in the women with lipoedema as significantly lower than the average for women in the Netherlands.

In an online survey of 120 women, Dudek et al (2016) identified higher quality of life in women with lipoedema as being correlated with higher levels of psychological flexibility and social connectedness. A further online survey by the same author (Dudek et al 2018) included 328 women from several countries including USA, UK, Netherlands, Germany Sweden and Australia, combining a variety of questionnaires to assess quality of life, symptom severity, mobility, appearance -related distress and depression. Using descriptive statistics, quality of life was reported as inversely related to lipoedema symptom severity, depression severity, and appearance-related distress. The authors surmised that distress related to appearance may affect eating, physical activity and lead to depression, anxiety, eating disorders and social isolation, suggesting that these issues should be addressed as part of treatment planning.

Experiences shared in online groups, such as the Talk Lipoedema Facebook groups, indicates that many women also experience significant distress associated with reproductive health issues, particularly when healthcare professionals do not understand lipoedema (see *Patient stories* box).

Management of lipoedema

Treatment approaches for lipoedema are limited in scope and availability, and there is no pharmacological or curative option presently available for women with lipoedema. As such, self-management is key, and includes healthy lifestyle, healthy eating, weight management, skin care, movement and physical exercise, alongside use of compression therapy. A Best Practice document developed in the UK (Wounds UK 2017) has identified the principles of lipoedema as:

- Supporting the person's ability to self-care including maintaining a healthy lifestyle
- Managing symptoms
- Optimising their health and preventing progression of the condition.

Further key considerations for managing and supporting women with lipoedema are outlined in Table 3.

Table 3: Managing and supporting women with lipoedema (Wounds UK, 2017)

- Listen to their story and support them to identify realistic goals
- Consider referral to appropriate services for example: lymphoedema services, psychological services, dieticians, physiotherapy, dermatology and wound care specialists
- Prescribe compression therapy where appropriate
- Prescribe analgesia where appropriate
- Avoid invasive or painful procedures such as venepuncture or taking blood pressure on an enlarged and painful lipoedematous limb
- Consider women's health issues that may cause significant problems:
    - Discuss contraceptive options with the women as some may exacerbate the lipoedema fat
    - Recognise the lipoedema may develop at puberty and worsen at menopause
    - Be aware that lipoedema may worsen significantly during pregnancy
    - Do not assume that the pregnant woman with lipoedema is unhealthy or 'obese' and be sensitive to the challenges during pregnancy and labour
    - Be prepared to discuss sexual difficulties including those related to poor mobility and high body weight
    - Recognise that some women with lipoedema may find it difficult to lift their children due to painful limbs.

Self-management

Working in partnership with the person and their significant family and carers will help to establish realistic expectations and goals for self-management (Fetzer & Wise 2015). Support with pain management is imperative, particularly to improve mobility and reduce social isolation. Low impact exercise including Pilates, yoga, swimming and Tai Chi are useful and well tolerated. Women with late stage lipoedema may require significant help with care and home adaptations, to ensure they remain as mobile as possible within and outside the home. Enabling individuals to effectively manage stress, psychological distress and anxiety associated with lipoedema is important as these may compromise their ability to self-care (Wounds UK 2017).

Dietary advice for women with lipoedema does not always mirror the NHS 'Eatwell Plate' and traditional weight-loss diets can have a disproportionate influence on non-lipoedema fat, further enhancing the lipoedema shape (Smith et al 2018). Many women with lipoedema find success in following an anti-inflammatory diet, avoiding a diet high in carbohydrate, sugar and processed foods, or a ketogenic diet with low carbohydrate and high (healthy) fats. This enables them to lose non-lipoedema fat and prepare themselves for treatments such as liposuction. Sadly, in the author's clinical experience, some women with lipoedema report health professionals as overly challenging, disbelieving their attempts to adhere to a healthy diet. Listening to individuals is a key message.

Treatments

The most common approaches to treatment include compression therapy, manual lymphatic drainage and liposuction (Wounds UK 2017).

<u>Compression therapy</u>: this includes prescription of compression garments, adjustable compression wraps and/or compression bandaging which contain the limb, buttocks and/or abdomen, and occasionally, the arms. The main aim is to reduce discomfort and pain by supporting the tissues, streamlining the uneven, distorted limb shape, improving mobility, and reducing oedema by encouraging venous and lymphatic return (Wounds UK 2017). Flat-knit, custom-made garments are widely available on prescription and may fit more comfortably than highly elastic stockings used, for example, in venous disease. As the feet may not be swollen in

lipoedema, footless legging garments are sometimes used. Measurement and fitting by a suitably qualified practitioner are required.

Manual lymphatic drainage (MLD): this gentle but specific type of massage will enhance lymphatic flow and may be helpful in managing symptoms of pain and oedema. This may be available in lymphoedema clinics or through private MLD therapists.

Liposuction: this is not widely available in the NHS and many women seek liposuction privately from clinics in the UK and specialist centres in Germany. The aim of liposuction is to reduce tissue bulk, pain and bruising, and by reducing the size of fat pads and tissue weight, improve mobility. There is evidence that liposuction can improve quality of life (Baumgartner et al 2016). Pre-operative counselling will ensure that the woman is realistic about what can be achieved and prepared for wearing compression garments post-operatively. Many women undergo several operations over time and despite the pain and obvious challenges of having surgery, many find it helpful in improving symptoms, although this is not a cure.

Patient stories
'Alison' was diagnosed with lipoedema one year ago. She has two children aged two and four. She described many difficulties during her pregnancies, when she did not yet have a diagnosis of lipoedema. Instead she was diagnosed as being 'obese'. During the pregnancy she had symptoms (which turned out to be severe cholecystitis) but were wrongly identified as chest pain due to what the hospital staff identified as her 'poor lifestyle'. Alison believes that the health professionals did not listen to her and made assumptions about her health and the health of her unborn child. This led to anxiety and depression and PTSD.
'Barbara' took herself off the contraceptive pill when she felt that it was negatively affecting her lipoedema. A few years later, when she was unable to get pregnant, she was refused fertility treatment due to her weight. She was unable to loose the lipoedema weight, and was told that her BMI was too high to meet the criteria for IVF.

‘Clare’ says: ‘*I can remember going to the midwife after my last child and we were talking about her being premature. The midwife said ‘She is a whopper isn’t she even though she is premature….how did you fit her in there as you are quite small…well in height ?’. She then glanced at my legs and rolled her eyes; my husband was there and he was as shocked as me’*.

‘Debs’ had symptoms of lipoedema since puberty. She tried to keep her legs hidden through her early years and battled with her weight, trying many different diets. At her first pregnancy when she was 26 she weighed 10 stone, but during her second pregnancy when she was 30, she gained another 10 stone as a result of the lipoedema. She worked hard to eat healthily and exercise regularly, and reduced to 15 stone. Ten years ago she came across a magazine article about lipoedema and seeing a picture, recognised her legs and put a name to her condition. Since then she has had three liposuction operations, her mobility has improved, pain and reduced and she is now waiting to see what changes occur in her menopause.

Conclusion

Lipoedema is a complex, long term and poorly understood fat disorder, that may have a significant impact on the lives of women and their families. Accurate and timely diagnosis of lipoedema ensures that women can access appropriate support and treatment. Informed self-management support is key to providing hope, improving quality of life, and enabling women to adopt healthy lifestyles. Living well within their family life and preventing lipoedema complications such as cellulitis and poor mobility will also minimise progression of lipoedema and avoid future dependence on NHS services.

Box 1: Further contacts

- Talk Lipoedema: www.talklipoedema.org
- Lipoedema UK: www.lipoedema.co.uk
- MLD UK:

- RCGP course available at: http://www.lipoedema.co.uk/royal-college-of-general-practitioners-elearning-course-on-lipoedema/ This e-learning course was developed in partnership with Lipoedema UK and is available free (Fetzer & Fetzer 2015).

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